SINONASAL ANEURYSMAL BONE CYST: ARTICLE REVIEW

AGOSTINO SERRA, ALESSANDRO GULINO, MILENA DI LUCA, ANGELO CONTI, ANTONINO MANIACI, GIANLUCA CAMPIONE, FRANCESCA MIGLIORE, VERONICA CASTRO, LUIGI MAIOLINO
Department G.F. Ingrassia, ENT Section, University of Catania, Catania, Italy

ABSTRACT

Aneurysmal Bone Cyst (ABC) are extremely rare in the head and neck region and even rare in sinuses. It's a benign multicystic mass that is locally-destructive and rapidly expandable. The etiopathogenesis, even today, is not entirely clear. However, hemorragic fluid content and septated appearance are the characteristic feature of ABC. The clinical presentation produces symptoms due to the compression of adjacent structures or as a result of a pathological fracture and depends on localization. Most common location in this area are the mandible and maxillary bones. The complete surgical excision is the treatment of choice. There are limited reports in literature on these tumors at the sinonasal level. We present a complete review of the literature as well as an unusual localization of ABC in this district.

Keywords*: aneurysmal bone cyst, paranasal sinuses, orbit, CT and MRI, Fess.*

DOI: 10.19193/0393-6384_2017_5_129

Received *November 30, 2016;* ***Accepted*** *May 20, 2017*

Introduction

The AneurysmaL bone cyst (ABC) is a benign bone lesion, apparently similar to non-neoplastic tumor, which expands from the bone to the surrounding tissue. It was identified in 1942 by Jaffe and Lichtenstein as a "blood-containing osseous tumor" and it owes its denomination "aneurysmal" to the morpho-radiological similarity with the "aneurysm"[1].

It prevalently affects long and short bones, the pelvis and the vertebral bodies; but in the literature, though, some insurgent cases in soft tissues are also described[2]. Only 2% of ABC occur in the head and neck region, predominantly affecting the jaw and the cheekbone and occasional reports have described it in the paranasal sinuses[3]. In these localizations ABCs can be locally destructive.

We present this review as well as an unusual localization of ABC at the nasosinusal level. We also attempt to clinically and therapeutically codify this clinical entity in the light of the numerous case reports in the literature.

Epidemiology

ABCs represent 1-2% of all primitive bone tumors with an annual incidence among the general population of the primitive form of 0.14/100,000 inhabitants[3]. It occurs at a young age, generally in the range 11 months to 20 years (mean age 11.6 years)[4-5] with a slight prevalence in females compared to men (F: M = 2: 1)[6].

Localization

Lesions involving the skull are rare, comprising only 3-6% of all ABCs[7]. ABCs have been reported to involve both the neurocranium (ethmoidal, sphenoidal, temporal, occipital, parietal and frontal) and viscerocranium (mandible, maxilla and zygoma)[8].

In a large series, Ariel et al. reported 238 cases of ABC, in which less than 5% were located on the skull and face, and the most common location in this area was the mandible and maxillary bones[9]. Only few cases of ABC have been found to involve the ethmoid bone in the entire body of medical literature (Fig.1).

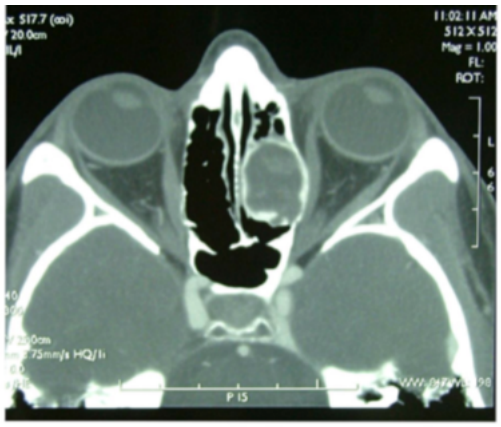

Figure 1: Ethmoid ABC treated with FESS in ENT Clinic of the University of Catania.

Etiopathogenesis

ABC is a benign bone lesion, the etiology of which is not entirely clear. There are limited reports on the pathophysiology and natural history of these tumors.

They are often classified as primary and secondary; with primary lesions appearing in isolation and secondary lesions appearing in the setting of another antecedent osseous lesion or as a result of trauma.

According to the “theory of hemodynamic disorder”, the pathogenesis of the lesion would be related to a sudden occlusion of the venous exhaust system with the consequent opening of artero-venous shunts, formation of blood filled spaces and subsequent suffering of bone tissue and trabecular thinning (Jaffè and Lichtenstein 1950).

Alternatively, it has been postulated that ABCs always arise as a secondary phenomenon in an antecedent lesion. The antecedent lesion undergoes involutional change, initiates an intraosseous arteriovenous malformation and thereby creates, via hemodynamic forces, a secondary bone reaction , which we know as an aneurysmal bone cyst[10].

Other authors found the presence of insulin-like growth factor (IGF-1) or specific sequences of mRNA coding for the same polypeptide in aneurysmatic bone cyst samples; this condition is apparently in contrast to normal bone tissue where IGF1 expression is extremely low[11].

The hypothesis of ABC's reactive nature has been questioned by the "genetic theory", first formulated by Panoutsakopoulos in a 1999 study, which demonstrated chromosomal translocation t (16; 17) (q22; p13) as a recurrent cytogenetic abnormality in ABC[12].

More recently, it has been demonstrated that t (16; 17) (q22; p13) induced the transfer and consequent fusion of the CDH11 promoter region (osteoblastic cadherin 11) located on chromosome 16q22 with the entire sequence of the USP6 gene (ubiquitin-specific protease 6 known as TRE2 or TRE17) located in chromosome 17p13, suggesting that pathogenesis of primary forms involves the upregolation of the USP6 transcript driven by the high activity of the CDH11 promoter region[13-15].

Clinical Aspects

The lesion often produces symptoms due to the compression of adjacent structures or as a result of a pathological fracture and depends on localization. Lesions involving the skull base are more likely to present with focal neurologic deficits. These can include anosmia visual deficits, facial numbness, ocular motility deficits and diplopia, hearing loss, facial weakness, jugular foramen syndrome, and ataxia. ABCs may also present with symptoms and signs of elevated intracranial pressure, spontaneous intracranial hemorrhage, seizure, obstructive hydrocephalus, proptosis, epistaxis and nasal obstruction, otalgia, otitis media, and ear mass[3,8,16-23].

Diagnosis

Histopathology

From a histologic point of view, it appears macroscopically as a circumscribed lesion, with dissections, which subdivides it into many intercommunicating cavernous spaces and with non-coagulated blood. Microscopically, a connective tissue-vascular tissue traced through various blood lacerations (sponge appearance) is highlighted (Fig.2). These blood gaps, because of their morphology, are considered pathognomonic of the pathology. Furthermore, within the lacuna spaces it is possible to highlight multinucleated giant cells, located around large vascular gaps and adhering to the walls. Two different types of ABC are considered: a typical variant (95% of the cases) and a solid variant (5%) that are indis-

tinguishable on clinical and radiological presentation and differ solely because the solid form does not present cavernous and sinusoidal spaces, but does present a greater number of mitotic figures and a definitive diagnosis of ABC depends on the pathological study of the specimen[1,4,6]. ABC may be difficult to differentiate, histologically, from giant cell tumor, fibrous dysplasia, ossifying hematoma and cavernous hemangioma of the bone due to the presence of multiple giant cells in all the above.

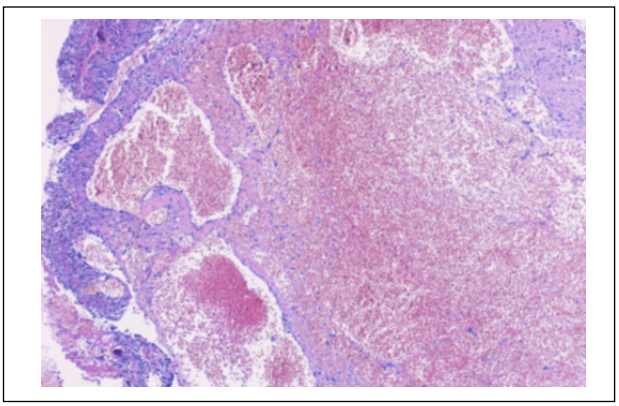

Figure 2: Vascular ectasia like aneurysmal (ENT Clinic of University of Catania).

Instrumental

Fundamental for differential diagnosis are CT and RI scans, as with these methods it is possible to highlight the extent of the pathology in soft tissues and the periosteum reaction.

In the CT examination, a hyperdense line is arranged to delimit the external contours of the lesion (cortical shell) and in some cases a cortical interruption may also be observed, although the periosteum remains intact. At MRI it appears as an expansive lesion with delimited margins and with the presence of sediments (honeycomb appearance), more evident after gadolinium enhancement. It is also possible to highlight the endocavitary fluid levels (characteristic but not a pathognomonic sign of this lesion), the presence of endocardial hemorrhages occurring in later periods (evidenced by the different signal intensity in the T1- and T2-weighed phases) and the presence of a peripheral orbit of a fibrous nature, separating the cyst from the adjacent tissues and evidenced as a low intensity signal in T1- and T2-weighed images, indicating the benignity of the neoformation and thus allowing differential diagnosis to be made with other malignant diseases such as osteogenic teleangectasis sarcoma. Preoperative arteriography highlights the vascularization of the lesion and thus makes pre-surgical embolization possible[3,8,24].

Differential diagnosis

These include giant cell tumor, giant cell reparative granuloma, hemorrhagic cyst, telangiectatic osteosarcoma, metastasis, plasmocytoma and also fibrous dysplasia. Giant cell tumors are usually seen in the older age group. Giant cell reparative granulomas usually have previous history of trauma, both of these present with hypointense signal in T2W and T1W images due to hamosiderin and/or fibrous components[25]. Fibrous dysplasia of the skull commonly involves the ethmoid sinuses and can diffuse, but it usually has a characteristic ground glass appearance on CT. The cortical margins, unlike in aneurismal bone cyst, are intact with minimal thinning and usually no fluid levels are seen[26-27]. Telangiectatic osteosarcoma may have radiological features similar to the aneurismal bone cyst but is more aggressive in appearance and the age group is older. On MRI, metastatic lesions show as a replacement of the normal marrow signal on T1 weighted images, they usually have a hyperintense signal on T2 weighted images and show enhancement of the bone and soft tissue component with gadolinium[28].

Condromixoid Fibroma is a benign neoplasm that is difficult to differentiate from ABC based only on radiological characteristics. Therefore it is essential and indispensable to perform a histological examination of the lesion, which, in the case of a condromixoid fibrous, will present a mixoid stroma with cartilage foci that are absent in the Aneurismatic Cyst[29].

Condroblastoma and Osteoblastoma are primitive bone lesions rarely associated with an aneurysmatic appearance, which makes the differential diagnosis quite easy. It is, however, desirable to emphasize a greater radiopacity of osteoblastoma due to the presence of ossified trabecules[30-31].

The epithelial adenomatoid hamartoma (REAH) of the nasal cavity is a polypoid proliferation of glandular spaces lined by ciliated epithelium and goblet cells in the upper aerodigestive tract[32].

Other sinonasal pathologies which may be related to ABC are inverted papilloma, non-ossifying fibroma, fibrous histiocytoma and low grade sinonasal carcinoma[3,33].

Treatment

In the treatment of the rhinobase tumors, complete surgical excision is the treatment of choice; it may range from conservative surgery such as curettage, enucleation and endoscopic surgery to more aggressive ones such as paralateral rhinotomy and bifrontal craniotomy for sinonasal ABC.

Author	Age (years), sex	Treatment
Baker et al. (1982)	20,F	Rinothomy
Jordan et al. (1983)	22,F	Rinothomy
Patel et al. (1993)	11,F	Rinothomy
Zielnik et al. (1995)	5,M	Rinothomy
Citardi et al. (1996)	12,M	Rinothomy
Winnepenninckx et al. (2001)	6,F	ESS
Hrischhikesh et al. (2002)	19,M	Bifrontal Craniotomy
Guilemany JM et al. (2004)	62,F	Rinothomy
Mazlout O et al. (2005)	15,F	Rinothomy
Ruiz de et al. (2007)	13,F	FESS
Serra A. et al (2007)	20,F	FESS
Cortese S. et al. (2008)	20,F	FESS
Bozbuğa M et al. (2009)	9,F	FESS
Wendt S et al. (2010)	3,F	FESS

Table 1: Surgical treatment
ESS: Endoscopic sinus surgery; FESS: Functional endoscopic sinus surgery

Endoscopic treatment provides a better visualization of the lesion, a reduced risk of postoperative morbidity, reduced hospitalization time, lower blood loss and, especially, the absence of an external incision; however, such a method is not free from contraindications such as size and location of the lesion and histological variation, therefore no external approach (external ethmoidectomy, maxillectomy or frontoethmoidectomy) can be ruled out as a possible alternative as it may be necessary in cases of underestimation of lesion staging and/or when they can be planned in association with the endoscopic approach in relation to the size of the lesion[34]. Selective arterial embolization in large lesions may help to improve surgery results. Radiotherapy is not recommended as several cases of evolution into sarcoma have been reported following this treatment[3,8] (Table 1).

The lesion recurrence rate within the first year after treatment is high and varies. The recurrence rate after resection is less than curettage. Recurrence rate of simple curettage varies from 21% to 50% and recurrence in radical methods varies from 11% to 25%. Further information about ethiopathogenesis and management of this entity is necessary[3,8,35-37].

Conclusions

ABC is a benign multicystic process that exceptionally appears in the nasal cavities, where only case reports are reported in the literature. The growth pattern is locally-destructive and rapidly expandable. Nevertheless, it should be taken into account when making a differential diagnosis. Clinical presentation depends on location, and lesions can often be identified preoperatively based on characteristic imaging findings. Gross total resection is the treatment of choice. Familiarity with skull base techniques is essential to resect lesions safely and completely arising in these locations.

References

1) Jaffè HL, Lichtenstein L. Solitary unicameral bone cyst, with emphasis on roentgen picture, pathologic appearance and pathogenesis. Arch Surg 44: 1004-1025, 1942.
2) Riccioni L, Foschini MP: Extraosseous aneurysmal bone cyst. Tumori 1996; 84: 485-487.
3) Hashemi SM, Heidarpour M, Eshaghian A, Ansari P, Hashemi MS, Yaghoobi M, Barati S. A Rare Case of Aneurysmal Bone Cyst in the Paranasal Sinus. Iran J Otorhinolaryngol. 2015 Sep; 27(82): 395-9.
4) Mankin H J, Hornicek F J, Ortiz-Cruz E, Villafuerte J, Gebhardt M C. Aneurysmal bone cyst: a review of 150 patients. J Clin Oncol. 2005; 23(27): 6756-6762.
5) Cuesta FR, Alenda C, Monerrisa E, et al. Aneurysmal bone cyst of the ethmoid: Apropos of a case and review of the medical literature. Acta Otorhinolaringol Esp. 2007; 58(3): 113-15.
6) Campidelli C , Di Tommaso L, Zanetti G. Aneurysmal bone cysts of the nasal cavity. Description of a case and review of the literature. Pathologica.2003 Apr; 95(2): 103-107.
7) Guida F, Rapanà A, Conti C, Cagliari E, Civelli F, Trincia G. Cranial aneurysmal bone cyst: a diagnostic problem. With a review of the literature. Childs Ner Syst 2001; 17(4-5): 297-301.
8) Hnenny L, Roundy N, Zherebitskiy V, Grafe M, Mansoor A, Dogan A. Giant Aneurysmal Bone Cyst of the Anterior Cranial Fossa and Paranasal Sinuses Presenting in Pregnancy: Case Report and Literature Review. J Neurol Surg Rep. 2015 Nov; 76(2): e216-21.
9) Ariel M, de Dios V, Bond JR, Shives TC, McLeod RA, Unni KK. Aneurysmal bone cyst, a clinicopathologic

study of 238 cases. Cancer. 1992: 69: 2921-31.
10) Biesecker JL, Marcove RC, HuvosAG, Miké V. Aneurysmal bone cysts. A clinicopathologic study of 66 cases. Cancer 1970; 26(3): 615-625.
11) Leithner A, Lang S, Windhager R, Leithner K, Karlic H, Kotz R, Haas OA. Expression of insulin-like growth factor-I (IGF-I) in aneurysmal bone cyst. Mod Pathol. 2001 Nov; 14(11): 1100-4.
12) Panoutsakopoulos G, Pandis N, Kyriazoglou I, Gustafson P, Mertens F. Recurrent t(16;17)(q22;p13) in aneurysmal bone cysts. Genes Chromosomes Cancer 1999; 26: 265-266.
13) Oliveira AM, Perez-Atayde AR, Inwards CY, Medeiros F, Derr V, Hsi BL, Gebhardt MC, Rosenberg AE, Fletcher JA.USP6 and CDH11 oncogenes identify the neoplastic cell in primary aneurysmal bone cysts and are absent in so-called secondary aneurysmal bone cysts. Am J Pathol. 2004 Nov; 165(5): 1773-80.
14) Oliveira AM, Perez-Atayde AR, Dal Cin P, Gebhardt MC, Chen CJ, Neff JR, Demetri GD, Rosenberg AE, Bridge JA, Fletcher JA. Aneurysmal bone cyst variant translocations upregulate USP6 transcription by promoter swapping with the ZNF9, COL1A1, TRAP150, and OMD genes. Oncogene. 2005 May 12; 24(21): 3419-26.
15) Guseva NV, Jaber O, Tanas MR, Stence AA, Sompallae R, Schade J, Fillman AN, Miller BJ, Bossler AD, Ma D. Anchored multiplex PCR for targeted next-generation sequencing reveals recurrent and novel USP6 fusions and upregulation of USP6 expression in aneurysmal bone cyst. Genes Chromosomes Cancer 2016 Nov 7.
16) Bathla G, Chowdhury V, Dixit R, Kottiyath VC, Jain R. Aneurysmal bone cyst of ethmoid sinus: Uncommon manifestation of a rare case. European Journal of Radiology Extra 70 (2009) e49-e51.
17) Serra A, Cocuzza S, Poli G, La Mantia I, Messina A, Pavone P. Otologic findings in children with gastroesophageal reflux. Int J Pediatr Otorhinolaryngol. 2007 Nov; 71(11): 1693-7. Epub 2007 Aug 22.
18) Matin N, Tabatabaie O, Falsaperla R, Lubrano R, et al. Epilepsy and innate immune system: A possible immunogenic predisposition and related therapeutic implications. Hum Vaccin Immunother. 2015; 11(8): 2021-9.
19) Serra A, Di Mauro P, Cocuzza S, Chiaramonte I, Maiolino L. Audiological findings in a case of cerebellar angioreticuloma. Acta Otorhinolaryngol Ital. 2015 Jun; 35(3): 212-4.
20) Janjua N, Cresswell M, Sharma R, Maheshwar A. Aneurysmal bone cyst of the ethmoid bone. BMJ Case Rep. 2014 Apr 4; 2014.
21) Serra A, Cocuzza S, Caruso E, Mancuso M, La Mantia I,. Audiological range in turner's syndrome. Int. J. Pediatric. Otorhinolaryngol. 2003 Aug; 67(8): 841-5.
22) Guilemany J.M., Alobid I., Blanch J.L., Ballesteros F., Alòs L., Mullol J. Orbitoethmoid aneurysmal bone cyst. Case report and literature review. Rhinology 2004 Sep; 42(3): 164-6.
23) Vitaliti G, Tabatabaie O, Matin N, Ledda C, et al. The usefulness of immunotherapy in pediatric neurodegenerative disorders: A systematic review of literature data. Hum Vaccin Immunother. 2015;1 1(12): 2749-63.
24) Sinha NR, Szmigielski W, Khanna M. Aneurysmal bone cyst of the ethmoid sinus: A case report. Pol J Radiol. 2010 Jul; 75(3): 65-7.
25) Bhalodiya NH, Singh N. Giant cell separative granuloma of posterior ethmoid: a case report. Ind. J. Otor. and Head and Neck Surg. Vol 57 No.4 2005; 325-327.
26) Dani RD, Gandhi VS, Thakkar GN, Prajapati HJ, Rathod GD. An Aneurysmal Bone Cyst Arising From Fibrous Dysplasia In Frontal Bone. Ind. J. Radiol. Imag. 2006; 16: 1: 49-52.
27) Terkawi A S, Al-Qahtani K H, Baksh E, Soualmi L, Mohamed Ael-B, Sabbagh A J. Fibrous dysplasia and aneurysmal bone cyst of the skull base presenting with blindness: a report of a rare locally aggressive example. Head Neck Oncol. 2011; 3: 15.
28) Murphey MD, Wan Jaovisidha S., Temple HT, Gannon FH, Jelinek JS, Malawer MM. Teleangiectatic osteosarcoma: radiologic-pathologic comparison. Radiology 2003 Nov; 229(2): 545-53.
29) Morris LG, Rihani J, Lebowitz RA, Wang BY. Chondromyxoid Fibroma of Sphenoid Sinus with Unusual Calcifications: Case Report with Literature Review. Head Neck Pathol. 2009 Jun; 3(2): 169-173.
30) Yong-Koo Park & Eui Jong Kim & Sung Wan Kim. Osteoblastoma of the ethmoid sinus. Skeletal Radiol.(2007); 36: 463-467.
31) Caltabiano R, Serra A, Bonfiglio M, Platania N, Albanese V, Lanzafame S, Cocuzza S. A rare location of benign osteoblastoma: case study and a review of the literature. Eur Rev Med Pharmacol Sci. 2012 Nov; 16(13): 1891-4.
32) Caltabiano R, Cocuzza S, Maira S, Gurrera A, Serra A, Lanzafame S. Respiratory epithelial adenomatoid hamartoma: case report and literature review. Pathologica. 2008 Jun; 100(3): 185-8.
33) Serra A, Caltabiano R, Spinato G, et al. Expression pattern of estroprogestinic receptors in sinonasal inverted papilloma. Oncotarget 2017 Apr. 17. doi:10.18632/oncotarget 17161.
34) Tirelli G, Gatto A, Spinato G, Tofanelli M. Surgical treatment of nasal polyposis: a comparison between cutting forceps and microdebrider. Am J Rhinol Allergy. 2013 Nov-Dec; 27(6): e202-6.
35) Vitaliti G, Pavone P, Matin N, Tabatabaie O, et al. Therapeutic approaches to pediatric pseudotumor cerebri: New insights from literature data. Int J Immunopathol Pharmacol. 2017 Mar; 30(1): 94-97.
36) Rapisarda V, Ledda C, Ferrante M, Fiore M, et al. Blood pressure and occupational exposure to noise and (LEAD Pb): a cross-sectional study. Toxicol. Ind Health, 2016 oct; 32(10): 1729-36.
37) Boscolo-Rizzo P, Tirelli G, Mantovani M, Baggio V, et al. Non endemic locoregionally advanced nasopharyngeal carcinoma: long-term outcome after induction plus concurrent chemoradiotherapy in everday clinical practice Eur Arch Otorhinolaryngol. 2015 nov; 272(11): 3491-8.

Corresponding author
LUIGI MAIOLINO
Department G.F. Ingrassia, ENT Section, University of Catania, Catania
(Italy)